ETHIOPIAN INTERNATIONAL JOURNAL OF MULTIDISCIPLINARY RESEARCH

eISSN: 2349-5715 pISSN: 2349-5707

Volume: 11, Issue 01, Jan-2024 SJIF 2019: 4.702 2020: 4.737 2021: 5.071 2022: 4.919 2023: 6.980

OPTICONEUROMYELITIS SPECTRUM DISEASE: A LITERARY REVIEW

Abdukadirova D.T.

Candidate of Medical Sciences Associate Professor

Kamalova N.L.

Department of Neurology assistant, ASMI.

Annotation: An analytical review of modern literature on epidemiology, pathogenesis, pathomorphology, clinic, diagnosis and treatment of Devik's disease (opticoneuromyelitis) is presented. The characteristic neurological symptoms of spinal cord and optic nerve damage are summarized. The diagnostic criteria of Devik's disease and opticomyelitis-associated disorders are presented. Spinal cord MRI disorders typical of Devic's disease have been studied. The high diagnostic significance of the antibody titer to aquaporin-4 has been shown. Promising directions are presented prevention of relapses with monoclonal antibodies.

Keywords: Devik's disease; antibodies to aquaporin-4; literature review, optic nerve damage, spinal cord damage.

Devik's disease (BD, optical neuromyelitis, G36.0 according to ICD-10) in the form of simultaneous or sequential development of optic neuritis and transverse myelitis was first described by the French physician E. Device in 1894 [9] and many years later received an eponymous name. For many years, this disease was considered one of the atypical clinical forms of multiple sclerosis, but the discovery in 2004 by V. Lennon et al. serum antibodies to aquaporin-4 (neuromyelitisoptica) specific for this pathology — Immunoglobulin G, NMO-IgG) suggested its independence [12, 13]. This pathology is one of the rare diseases, but it is more common among people of African and Asian descent [14]. In European countries, the prevalence of DB in epidemiological studies is 0.3–4.4 cases per 100,000 population [8, 11]. The age of onset of the disease varies in a wide range with a peak of onset in 35-41 years. Women suffer from DB significantly more often (85% of cases) than men. Despite the 125-year history of studying this diseases in Russia and Ukraine, to date, only a few scientific articles have been published describing single observations of DB [1-4, 7]. At the same time, the largest series of cases of DB diagnosis (30 people) was described by T.O. Simaniv in 2015 [6].

It has been established that the development of this pathology of the central nervous system is based on an autoimmune process involving cellular and humoral links of immunity. The high titer of NMO-IgG, which is part of the membrane of the macromolecular complex of astrocytes as part of the blood-brain barrier, plays a leading role in confirming violations of the humoral mechanisms of immunity in DB. Its defeat causes the opening of water channels, leads to the loss of intracellular Na^+ and the ingestion of glutamate into neurons, which causes their apoptosis. According to T. Misuetal. The sensitivity of NMO-IgG detection in DB is 91%, and the specificity reaches 100%. In addition, it has been shown that the titer of NMO-IgG in blood serum correlates with the size of foci in the spinal cord and the frequency of DB exacerbations. Violation of cellular mechanisms of water transport, damage to the blood-brain barrier and infiltration of the perivascular space by cellular elements of the blood (neutrophils, macrophages) contribute to the process of demyelination in white and gray matter mainly of the spinal cord, as well as optic nerves. Thus, The pathogenesis of DB differs from multiple sclerosis autoimmune astrocytopathy and/or channelopathy, which has a predominantly humorally mediated mechanism [5].

eISSN: 2349-5715 pISSN: 2349-5707
Volume: 11, Issue 01, Jan-2024 SJIF 2019: 4.702 2020: 4.737 2021: 5.071 2022: 4.919 2023: 6.980

The disease begins subacute or has a chronic course. In half of the cases, the development of BD is preceded by previous infections or stress, in the rest it begins for no apparent reason [1, 5]. Patients (up to 90%) with a recurrent course predominate [6]. A monophasic course is less common (15-25%); at the same time, a secondary progressive course is not typical for DB. Predictive The factors of the recurrent course of the disease are short remissions (up to 6 months), female sex, late age of onset, residual motor deficiency after exacerbation of the process, the presence of concomitant autoimmune pathology and the presence of NMO-IgG in serum [10].

Pathomorphologically, DD affects the optic nerves, chiasm, gray and white matter of the spinal cord, less often the hypothalamus and brain stem. Microscopy reveals inflammatory infiltration, a focus (foci) of complete necrosis with lipophages, axonal balls, a zone of demyelination around the focus, lymphocytic and plasmocytic perivascular infiltrates in the spinal cord, hemispheres of the cerebrum and its trunk, hypertrophy and proliferation of astrocytes, microgliocytes. In the optic nerve as well foci of demyelination, lymphocytic infiltration and hypertrophied astrocytes [1].

Neurological disorders in DD are associated with predominant damage to the spinal cord and optic nerves. More often, the cervical and thoracic parts of the spinal cord suffer from this pathology. The clinical signs of DD depend on the prevalence of the demyelinating process along the length and diameter of the spinal cord, the level of lesion and range from pyramidal insufficiency to symmetrical para- or tetraplegia (66%). Sensitive disorders are characteristic conduction type and pelvic (58%) disorders. Neuropathic pain in the extremities (33%), Lermitt's symptom and painful cramps (cramps) in the legs are common [8]. After an acute episode of myelitis in BD, only partial recovery of motor functions is usually observed, while a complete regression of all neurological symptoms, unlike multiple sclerosis, is not characteristic [6].

In cases of the spread of the demyelinating process to the medulla oblongata, uncapped hiccups, vomiting, and non-systemic dizziness, nystagmus and neuroendocrine disorders (amenorrhea, galactorrhea) [5]. Some patients with DD have dry mouth due to concomitant inflammatory infiltration of the salivary glands.

Damage to the optic nerves in DD more often (up to 80%) may precede damage to the spinal cord or occur simultaneously with it, or join later, after several months or years. Moreover, visual symptoms can vary from subclinical in the form of paleness of the optic nerve discs, which are detected only with a targeted examination by an optometrist, up to of a pronounced degree with the development of blindness following primary atrophy. Neuritis of the optic nerves in DB is more often bilateral (41%), less often unilateral (20%). Positive visual symptoms are characteristic in the form of pain in the orbit, flickering lights, spots, lines, changes in color perception [1-4, 7]. During retinal papillarometry in patients with DB, thinning of optic nerve fibers is detected, the severity of which correlates with visual acuity, contrast vision scales and the number of relapses.

In the diagnosis of DD, in addition to assessing the clinical picture, the study of cerebrospinal fluid (CSF) and conducting MRI of the brain and spinal cord. CSF in the acute phase of the disease is characterized by the appearance of insignificant (no more than 50 cells /mm3)) lymphocytic pleiocytosis. Oligoclonal antibodies characteristic of multiple sclerosis in CSF are not typical for DD [5].

A characteristic MRI sign of DD is the detection of T2W-mode of hyperintensive foci (or merging foci of demyelination) with involvement both gray and white matter of the spinal cord, elongated in the cranio-caudal direction, with a length of at least three vertebral segments. These foci are capable

eISSN: 2349-5715 pISSN: 2349-5707
Volume: 11, Issue 01, Jan-2024 SJIF 2019: 4.702 2020: 4.737 2021: 5.071 2022: 4.919 2023: 6.980

of accumulating contrast, and rarely have a mass effect. The absence of foci characteristic of multiple sclerosis is diagnostically significant during simultaneous MRI of the brain.

The diagnostic criteria of the database are presented in Table 1.

Criteria	Diagnostic signs
I. Absolute	1. Optical neuritis. 2. Transverse myelitis. 3. The absence of other diseases with this symptom
II. Large supportive	1. The absence of signs of multiple sclerosis in MRI of the brain. 2. During MRI of the spinal cord, the detection of T2W hearth, exciting three or more vertebral segment. 3. Detection of lymphocytic pleocytosis in CSF > 50 cells/mm3 or > 5 neutrophils/mm3
III. Small supportive	1. Bilateral optic neuritis. 2. Pronounced optical neuritis with stable visual impairment less than less than 20/200 for at least one eye. 3. Pronounced stable weakness in the extremities at the time of exacerbation with a decrease in strength of at least 2 points in one or more extremities

When making a diagnosis, it is important to consider. The criteria for excluding DD are "red flags", which include clinical, laboratory data and MRI results of spinal cord examination.

1. Clinical features or laboratory results confirming the diagnosis of multiple sclerosis:

— atypical time of exacerbation (deterioration less than 4 hours, the duration of exacerbation is more than 4 weeks);

— partial myelitis with an unusual pattern for DD in MRI of the spinal cord;

— detection of oligoclonal antibodies in CSF.

2. The presence of comorbid diseases:

— sarcoidosis, lymphomas, paraneoplastic syndromes;

— chronic infections (HIV, syphilis).

3. MRI results of the brain and spinal cord:

— foci of demyelination in T2w with pen orientation pendicular to the corpus callosum ("Dawson's fingers");

— foci of demyelination directed from the lateral ventricles to the lower temporal lobes;

eISSN: 2349-5715 pISSN: 2349-5707
Volume: 11, Issue 01, Jan-2024 SJIF 2019: 4.702 2020: 4.737 2021: 5.071 2022: 4.919 2023: 6.980

— subcortical foci of demyelination involving U-fibers;

— the focus of demyelination in the spinal cord is less three vertebral segments.

In addition, it should be borne in mind that similar clinical symptoms can be detected in opticomyelitis-associated disorders, which are more often observed in the framework of systemic diseases [6].

Table 2. Diagnostic criteria for opticomyelitis-associated disorders with and without NMO-IgG

Diagnostic criteria for opticomyelitis-associated disorders with NMO-IgG	Diagnostic criteria for opticomyelitis-associated disorders in the absence or unknown status NMO-IgG
1. At least one main clinical symptom. 2. A positive test for NMO-IgG with the most informative of the existing antibody detection methods. 3. Exclusion of alternative diagnoses	1. At least 2 main clinical symptoms resulting from one or more clinical exacerbations and corresponding to all of the following characteristics: A. At least one exacerbation should relate to optic neuritis, acute longitudinal myelitis. B. Dissemination at the site (2 or more different clinical symptoms belonging to the main category). c. Compliance with the additional requirements for MRI. 2. Negative test for NMO-IgG using the most informative of the existing methods detection of antibodies or the inability to perform a test. 3. Exclusion of alternative diagnoses

Thus, BD and other opticomyelitis-associated disorders are among the rare clinical forms of demyelinating diseases of the central nervous system. Despite its relative rarity, this pathology has a well -defined, albeit polymorphic, clinical picture and strict diagnostic criteria. With timely diagnosis and adequate pathogenetic therapy, BD has a high 5-year survival rate and potential for recovery lost motor and visual functions.

Literature:

1. Gulevskaya T.S., Morgunov V.A., Erokhina L.G. and others. A case of a rare demyelinating disease — opticoneuromyelitis (Devik's disease): clinic and pathomorphology. Neurological journal. 2001. No. 1. pp. 25-30.

2. Wooden E.A., Shepilova I.D., Karamysheva K.N., etc. Devik's disease (opticomyelitis): a clinical case and review literatures. Bulletin of the Council of Young Scientists and Specialists Chelyabinsk region. 2016. No. 3. pp. 41-47.

eISSN: 2349-5715 pISSN: 2349-5707

Volume: 11, Issue 01, Jan-2024 SJIF 2019: 4.702 2020: 4.737 2021: 5.071 2022: 4.919 2023: 6.980

3. Isaikin A.I., Schmidt T.E., Yakhno N.N. et al. Opticoneuromyelitis. Neurological Journal. 2014. No. 5. pp. 45-51.

4. Mironenko T.V., Khubetova I.V. Opticomyelitis (disease Devika): scientific review and own clinical observation. International Journal of Neurology. 2015. No. 1.pp. 141-147.

5. Ponomarev V.V. Autoimmune diseases in neurology. Minsk. 2010. pp. 41-45.

6. Simaniv T.O. Opticomyelitis and opticomyelitis-associated syndromes in demyelinating diseases: abstract of the dissertation of the Candidate of Medical Sciences. Moscow. 2015.

7. Yakhno N.N., Mozolevsky Yu.V., Golubeva V.V. et al. Opticomyelitis of a girl. Neurological Journal. 2008. No. 2. pp. 27-32.

8. Rizvi S. Clinical Neuroimmunology. Multiple Sclerosis and Related Disorders. Humana Pres. 2012. P. 219-232.

9. Devic E. Myelitesubaiguecompliquee de neuriteoptique. Bull. Med. 1894. № 8. P. 1033-1034.

10. Jacob A., Weinshenker B.G., Violich I. et al. Treatment of neuromyelitisoptica with rituximab: retrospective analysis of 25 patients. Arch. Neurol. 2008. Vol. 65. № 11. P. 1443-1448.